The Cardiovascular System

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طب عام ج/ الكوفةَ

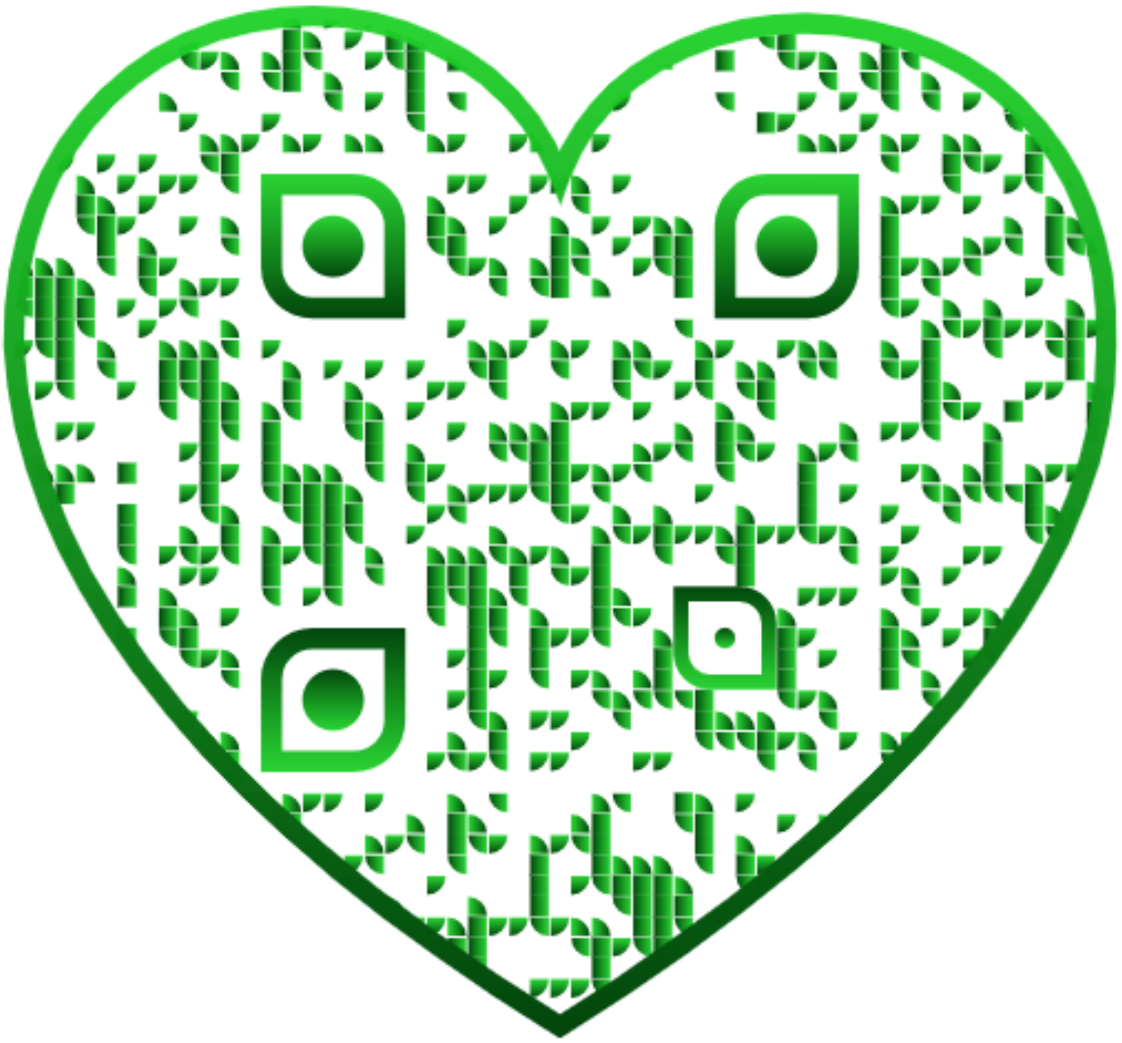
175. A 14-year-old girl, angry at her mother for taking away her MP3 player, takes an unknown quantity of a friend's pills. Within the first hour she is sleepy, but in the emergency center she develops a widened QRS complex on her electrocardiogram (ECG), hypotension, and right bundle branch block. The therapy you would initiate for this ingestion is which of the following?

a. *N*-acetylcysteine (Mucomyst)
b. Naloxone
c. Intensive care unit (ICU) admission, close monitoring, and possible Fab antibody fragments
d. Ethanol
e. Deferoxamine

The clinical presentation is that of a tricyclic antidepressant ingestion. In smaller children the central nervous system (CNS) symptoms of drowsiness, lethargy, coma, and seizures are more commonly seen than the cardiac effects of tachycardia, initial hypertension followed by hypotension, and a variety of cardiac effects such as widening of the QRS complex and bundle branch blocks which are often seen in adolescents. Close monitoring in the ICU and tricyclic antidepressant Fab antibody fragments (if available) are administered.

176. A 10-year-old boy, the star pitcher for the Salt Lake City Little League baseball team, had a sore throat about 2 weeks ago but did not tell anyone because he was afraid he would miss the play-offs. Since several children have been diagnosed with rheumatic fever in the area, his mother is worried that he may be at risk as well. You tell her that several criteria must be met to make the diagnosis but the most common finding is which of the following?

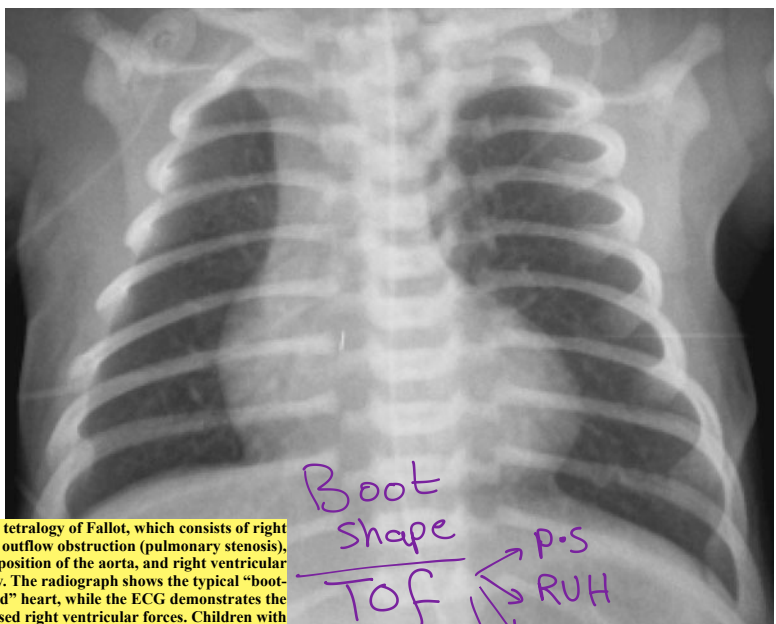
a. Carditis
b. Arthralgia
c. Erythema marginatum
d. Chorea
e. Subcutaneous nodules



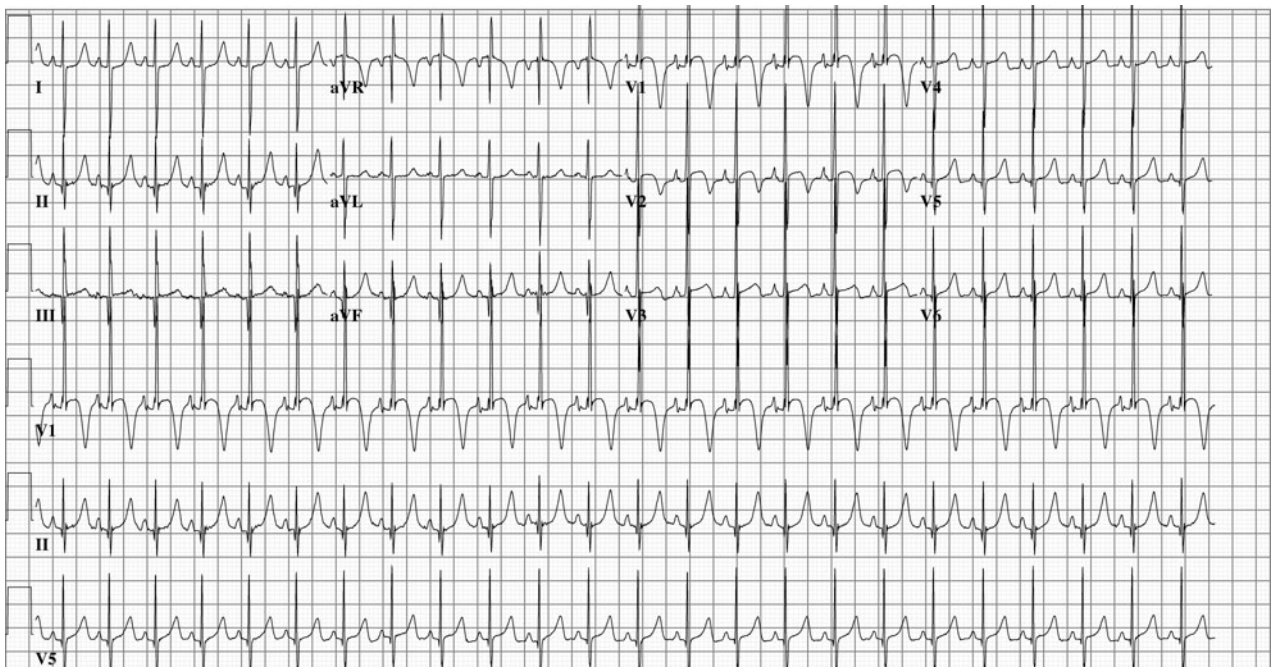
177. You are seeing a 2-year-old boy for the first time. His father denies any past medical or surgical history, but does note that the child's day care recently sent a note home asking about several episodes, usually after the child does not get what he wants, when he "breathes funny" and sits in a corner with his knees under his chin for a few minutes. The day-care staffers think this "self-imposed time-out" is a good thing, but they worry about the breathing. One teacher even thought he once looked blue, but decided that it was probably because of the finger paints he had been using. On examination, you identify a right ventricular impulse, a systolic thrill along the left sternal border, and a harsh systolic murmur (loudest at the left sternal border but radiating through the lung fields). His chest radiograph and ECG are shown. Which of the following congenital cardiac lesions would you expect to find in this child?

Knee Chest position

a. Patent ductus arteriosus
b. Right ventricular outflow obstruction
c. Atrial septal defect (ASD)
d. Transposition of the great vessels with a patent foramen ovale
e. Hypoplastic left heart



This child has tetralogy of Fallot, which consists of right ventricular outflow obstruction (pulmonary stenosis), VSD, dextroposition of the aorta, and right ventricular hypertrophy. The radiograph shows the typical "boot-shaped" heart, while the ECG demonstrates the increased right ventricular forces. Children with tetralogy of Fallot may have cyanotic episodes ("tet spells") associated with acute reduction in pulmonary blood flow. Typically, these spells are self-limited, lasting no more than 30 minutes. Assuming the knee-chest position is thought to increase peripheral resistance, decreasing the amount of right-to-left shunting and thus increasing pulmonary blood flow. Alternative therapies include morphine sulfate and propranolol. Prolonged hypoxia can lead to acidosis; correction may require infusion of sodium bicarbonate.



B

(Courtesy of Steven Lorch, MD.)

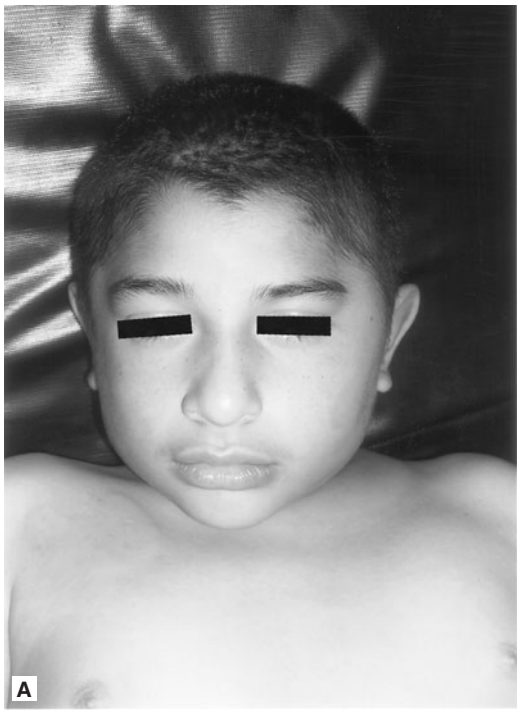
178. The parents of a 2-month-old baby boy are concerned about his risk of coronary artery disease because of the recent death of his 40-year-old maternal uncle from a myocardial infarction. Which of the following is the most appropriate management in this situation?

a. Screen the parents for total cholesterol.
b. Counsel the parents regarding appropriate dietary practices for a 2-month-old infant and test him for total cholesterol at 6 months of age.
c. Reduce the infant's dietary fat to less than 30% of his calories by giving him skim milk.
d. Initiate lipid-lowering agents.
e. Recommend yearly ECGs for the patient.

Children with a first- or second-degree relative with early onset of coronary heart disease should be evaluated early in life but after 2 years of age. Other known risk factors include obesity, diabetes, hypertension, and smoking. No change in current infant dietary practice is recommended for children under 2 years of age.

179. For the past year, a 12-year-old boy has had recurrent episodes of swelling of his hands and feet, which has been getting worse recently. These episodes occur following exercise and emotional stress, last for 2 to 3 days, and resolve spontaneously. The last episode was accompanied by abdominal pain, vomiting, and diarrhea. The results of routine laboratory workup are normal. An older sister and a maternal uncle have had similar episodes, but they were not given a diagnosis. He presents today with another episode as shown in the photographs on the next page. Which of the following is the most likely diagnosis?

a. Systemic lupus erythematosus
b. Focal glomerulosclerosis
c. Congenital nephrotic syndrome
d. Hereditary angioedema
e. Henoch-Schönlein purpura



(Courtesy of Adelaide Hebert, MD.)

180. A 15-year-old girl with short stature, neck webbing, and sexual infantilism is found to have coarctation of the aorta. A chromosomal analysis likely would demonstrate which of the following?

a. Mutation at chromosome 15q21.1
b. Trisomy 21
c. XO karyotype
d. Defect at chromosome 4p16
e. Normal chromosome analysis

181. A newborn is diagnosed with congenital heart disease. You counsel the family that the incidence of heart disease in future children is which of the following?

a. 1%
b. 2% to 6%
c. 8% to 10%
d. 15% to 20%
e. 25% to 30%

182. During a regular checkup of an 8-year-old child, you note a loud first heart sound with a fixed and widely split second heart sound at the upper left sternal border that does not change with respirations. The patient is otherwise active and healthy. Which of the following heart lesions most likely explains these findings?

a. Atrial septal defect (ASD)
b. Ventricular septal defect (VSD)
c. Isolated tricuspid regurgitation
d. Tetralogy of Fallot
e. Mitral valve prolapse

183. A 2-year-old boy is brought into the emergency room with a complaint of fever for 6 days and the development of a limp. On examination, he is found to have an erythematous macular exanthem over his body as shown in image A, ocular conjunctivitis, dry and cracked lips, a red throat, and cervical lymphadenopathy. There is a grade 2/6 vibratory systolic ejection murmur at the lower left sternal border. A white blood cell (WBC) count and differential show predominant neutrophils with increased platelets on smear. Later, he develops the findings as seen in image B. Which of the following is the most likely diagnosis?

a. Scarlet fever
b. Rheumatic fever
c. Kawasaki disease
d. Juvenile rheumatoid arthritis
e. Infectious mononucleosis

Kawasaki Disease (KD) : Vasculitis of medium and small-sized blood vessels

Diagnostic criteria

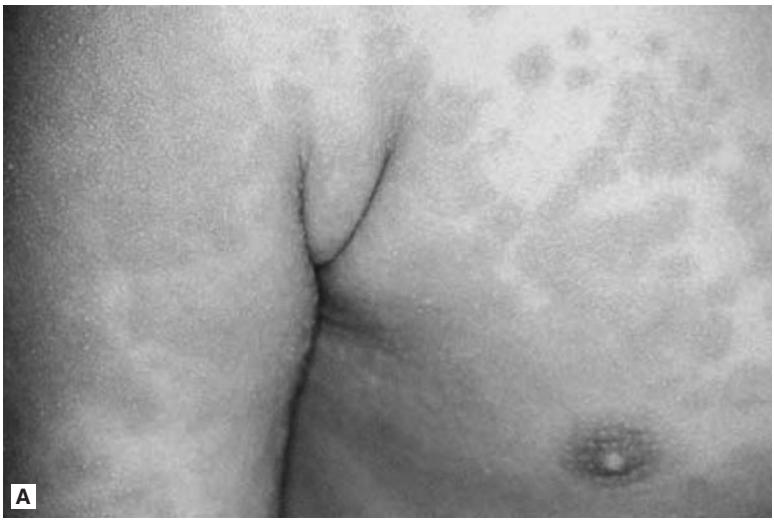
- Prolonged unexplained fever of >38. 5°C ≥ five days plus at least 4 out of:
  - Bilateral non exudative conjunctivitis
  - Mucositis: cracked, red lips , a strawberry tongue and injected pharynx
  - Polymorphous rash: perineal erythema, followed by macular, morbilliform, or targetoid skin rash of the trunk and extremities
  - Extremity changes: edema of the dorsum of hands and feet, and a diffuse erythema of palms and soles
  - Cervical lymphadenopathy; at least one lymph node >1.5 cm in diameter.
- KD carries risk of coronary aneurysms and infarction

Investigations

- Elevated acute phase reactants and thrombocytosis
- Echocardiography follow-up for coronary aneurysms

Treatment

- IVIG 2gm/kg IV infusion over 8-12 hours
- Aspirin oral 80-100 mg/kg till fever decline for 48 hours then antiplatelet dose 3-5mg/kg till acute phase reactants normalizes



(Reproduced with permission from Wolff K, Johnson RA, Suurmond D. Fitzpatick's Color Atlas & Synopsis of Clinical Dermatology. *5th ed. New York, NY: McGraw-Hill; 2005.)*

(Reproduced with permission from Wolff K, Goldsmith LA, Katz SI, et al. Fitzpatick's Dermatology in General Medicine. *7th ed. New York, NY: McGraw-Hill; 2007.)*

184. An ill-appearing 2-week-old baby girl is brought to the emergency room. She is pale and dyspneic with a respiratory rate of 80 breaths per minute. Heart rate is 195 beats per minute, heart sounds are distant, a gallop is heard, and she has cardiomegaly on x-ray. An echocardiogram demonstrates poor ventricular function, dilated ventricles, and dilation of the left atrium. An ECG shows ventricular depolarization complexes that have low voltage. Which of the following is the most likely diagnosis based on this clinical picture?

a. Myocarditis
b. Endocardial fibroelastosis
c. Pericarditis
d. Aberrant left coronary artery arising from pulmonary artery
e. Glycogen storage disease of the heart

The most common causes of myocarditis include adenovirus and coxsackievirus B,

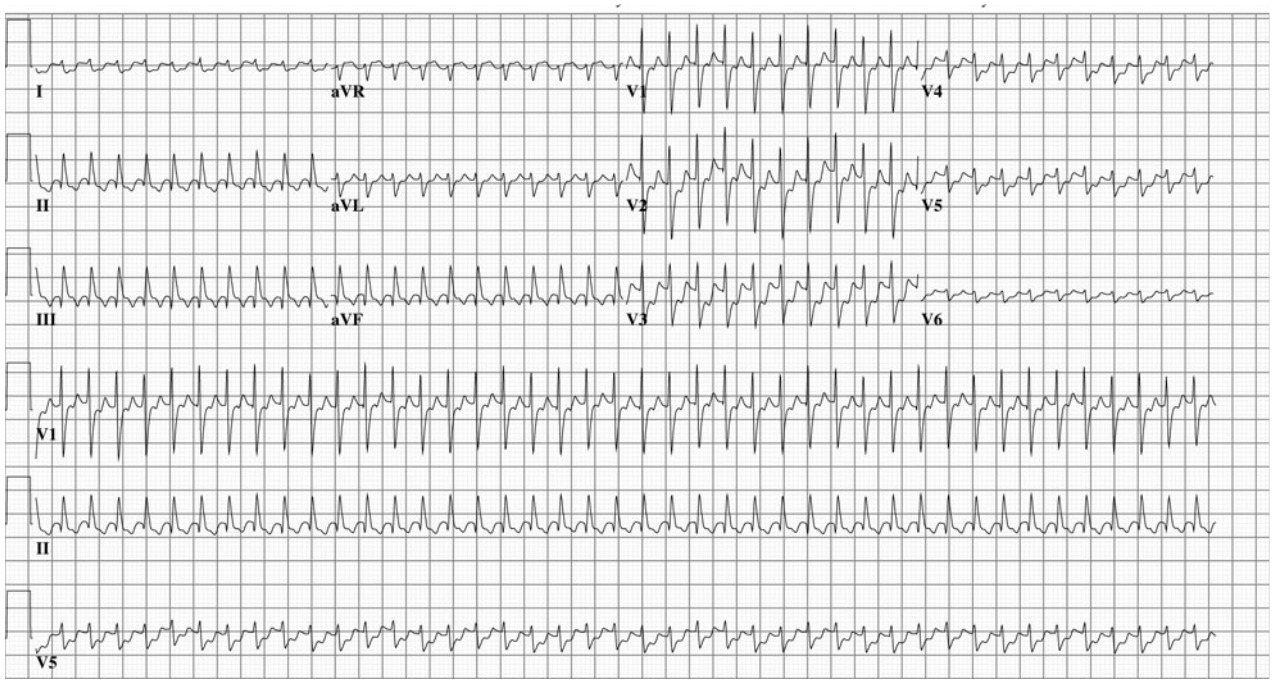
185. A newborn infant has mild cyanosis, diaphoresis, poor peripheral pulses, hepatomegaly, and cardiomegaly. Respiratory rate is 60 breaths per minute, and heart rate is 250 beats per minute. The child most likely has congestive heart failure caused by which of the following?

a. Large ASD and valvular pulmonic stenosis
b. VSD and transposition of the great vessels
c. Total anomalous pulmonary venous return
d. Hypoplastic left heart syndrome
e. Paroxysmal atrial tachycardia

Congestive heart failure from many causes can be associated with a rapid pulse rate (up to 200 beats per minute). A pulse rate greater than 250 beats per minute, however, should suggest the presence of a tachyarrhythmia. Common causes for supraventricular tachycardia include Wolff-ParkinsonWhite (WPW) syndrome, congenital heart disease, and sympathomimetic drugs

186. A 3-month-old infant is brought to your office for pallor and listlessness. Your physical examination reveals tachycardia that is constant and does not vary with crying. He has no hepatomegaly and the lungs are clear. His ECG is shown. Which of the following is the most appropriate initial management of this patient?

a. Rapid verapamil infusion
b. Transthoracic pacing of the heart
c. Carotid massage
d. DC cardioversion
e. Precordial thump



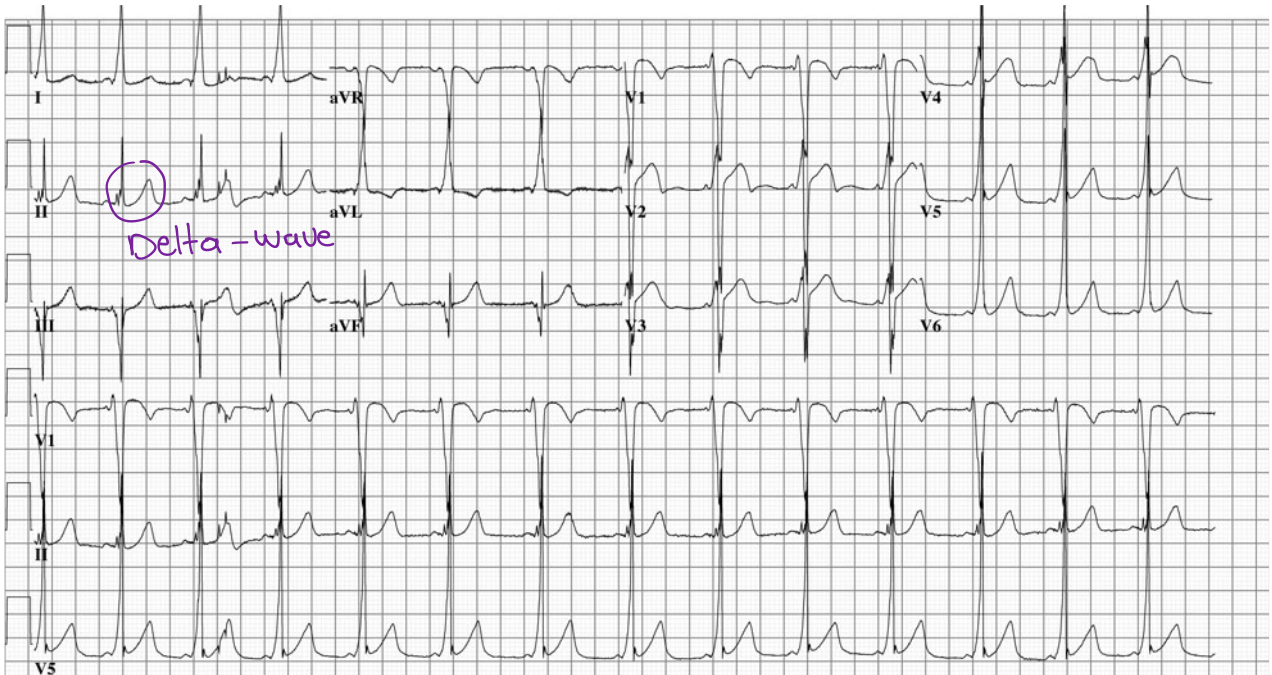
(Courtesy of Steven Lorch, MD.)

187. A 2-year-old child with minimal cyanosis has an S_3 and S_4 (a quadruple rhythm), a systolic murmur in the pulmonic area, and a middiastolic murmur along the lower left sternal border. An ECG shows right atrial hypertrophy and a ventricular block pattern in the right chest leads. Which of the following is the most likely diagnosis?

a. Tricuspid regurgitation and pulmonic stenosis
b. Pulmonic stenosis and a VSD (tetralogy of Fallot)
c. Atrioventricular canal
d. Ebstein anomaly ASD + Rt. to Lf. shunt
e. Wolff-Parkinson-White syndrome

188. A 4-year-old girl is brought to the pediatrician's office. Her father reports that she suddenly became pale and stopped running while he had been playfully chasing her and her pet Chihuahua. After 30 minutes, she was no longer pale and wanted to resume the game. She has never had a previous episode and has never been cyanotic. Her physical examination was normal, as were her chest x-ray and echocardiogram. An ECG showed the pattern seen on the next page, which indicates which of the following?

a. Paroxysmal ventricular tachycardia
b. Paroxysmal supraventricular tachycardia
c. Wolff-Parkinson-White syndrome
d. Stokes-Adams pattern
e. Excessive stress during play



(Courtesy of Steven Lorch, MD.)

189. A child has a 2-week history of spiking fevers, which have been as high as 40°C (104°F). She has spindle-shaped swelling of finger joints and complains of upper sternal pain. When she has fever, the parents note a faint salmon-colored rash that resolves with the resolution of the fever. She has had no conjunctivitis or mucositis, but her heart sounds are muffled and she has increased pulsus paradoxus. Which of the following is the most likely diagnosis?

a. Rheumatic fever
b. Juvenile rheumatoid arthritis
c. Toxic synovitis
d. Septic arthritis
e. Osteoarthritis

190. A cyanotic newborn is suspected of having congenital heart disease. He has an increased left ventricular impulse and a holosystolic murmur along the left sternal border. The ECG shows left-axis deviation and left ventricular hypertrophy (LVH). Which of the following is the most likely diagnosis?

a. Transposition of the great arteries
b. Truncus arteriosus
c. Tricuspid atresia
d. Tetralogy of Fallot
e. Persistent fetal circulation

Patients with tricuspid atresia typically have a hypoplastic right ventricle, and therefore the ECG shows left-axis deviation and left ventricular hypertrophy;

191. A 3-day-old infant with a single second heart sound has had progressively deepening cyanosis since birth but no respiratory distress. Chest radiography demonstrates no cardiomegaly and normal pulmonary vasculature. An ECG shows an axis of 120° and right ventricular prominence. Which of the following congenital cardiac malformations is most likely responsible for the cyanosis?

a. Tetralogy of Fallot → not early cyanosis
b. Transposition of the great vessels
c. Tricuspid atresia → early cyanosis bc2 Diminshed pulmonary flow
d. Pulmonary atresia with intact ventricular septum
e. Total anomalous pulmonary venous return below the diaphragm → snowman bc2 pulmonary venous congestion

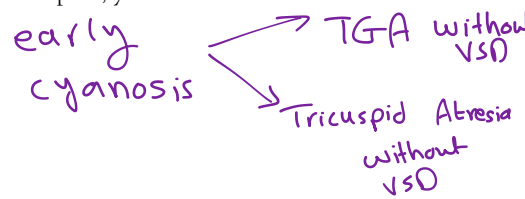
Transposition of the great vessels with an intact ventricular septum presents with early cyanosis, a normal-sized heart (classic "egg on a string" radiographic pattern in one-third of cases), normal or slightly increased pulmonary vascular markings, and an ECG showing right-axis deviation and right ventricular hypertrophy.

192. During a physical examination for participation in a sport, a 16-year-old girl is noted to have a late apical systolic murmur, which is preceded by a click. The rest of the cardiac examination is normal. She states that her mother also has some type of heart "murmur" but knows nothing else about it. Which of the following is the most likely diagnosis?

a. ASD
b. Aortic stenosis
c. Tricuspid regurgitation
d. Mitral valve prolapse
e. VSD

193. A previously normal newborn infant in a community hospital nursery is noted to be cyanotic at 14 hours of life. She is placed on a face mask with oxygen flowing at 10 L/min. She remains cyanotic, and her pulse oximetry reading does not change. An arterial blood gas shows her Pa_{O_2} to be 23 mm Hg. Bilateral breath sounds are present, and she has no murmur. She is breathing deeply and quickly, but she is not retracting. While you are waiting for the transport team from the nearby children's hospital, you should initiate which of the following?

a. Indomethacin infusion
b. Saline infusion
c. Adenosine infusion
d. Prostaglandin E_1 infusion
e. Digoxin infusion



Questions 194 to 196

For each case presented, select the cardiovascular defect with which the examination findings are most likely to be associated. Each lettered option may be used once, more than once, or not at all.

a. Coarctation of the aorta
b. ASD
c. Mitral valve prolapse
d. Pulmonic stenosis
e. Patent ductus arteriosis

The answers are 194-b, 195-d, 196-c.

b

194. An infant in the neonatal intensive care unit (NICU) has shortened lower distal arms but with thumbs. Her platelet count is profoundly low.

Thrombocytopeniaabsent radius syndrome (TAR)

d

195. A 15-year-old boy comes to your office for a Special Olympics sports physical. His height is in the 3rd to 5th percentile and his weight is in the 50th percentile. Physical examination reveals a young man with shieldlike chest, cryptorchidism, low-set and malformed ears, ptosis, and pectus excavatum.

Noonan Syn.

C

196. A new patient to your practice, a happy 10-year-old girl has a history of hyperextensible skin and easy bruising. She often has sprains and joint dislocations when she exercises.

Ehler Danlos syn.

Questions 197 to 200

For each of the cases presented below, choose the most likely cardiac diagnosis based on the patient's presentation. Each lettered option may be used once, more than once, or not at all.

a. Congenital heart block
b. Hypertrophic cardiomyopathy
c. Prolonged QT syndrome
d. Congestive heart failure
e. Cor pulmonale

b

197. A 15-year-old adolescent male presents to the office for a sports physical. In his screening questionnaire, he notes that he occasionally gets short of breath and dizzy during exercise, with occasional chest pain. He lost consciousness once last summer during football practice, but attributed it to the heat. His grandfather died suddenly at the age of 35 of unknown etiology, but otherwise the family is healthy.

C

198. A 15-year-old adolescent female comes to be evaluated for syncopal episodes. Her only other medical problem is congenital deafness. She notes through a signing interpreter that syncopal episodes happen during stressful or emotional situations and that they started only within the past year or so. Her mother and father are from the same small farming town and are second cousins.

199. You are called to the nursery to evaluate a newborn. The mother has a history of systemic lupus erythematosus and gestational diabetes. The nurses are concerned because the baby has developed petechiae and bruising after his bath. Vital signs have been stable, with a heart rate in the 60 beats per minute range and a respiratory rate in the 40 breaths per minute range. You note a large liver, scattered petechiae, and an erythematous rash on the cheeks and on the bridge of the nose.

A

200. An infant previously diagnosed with a large muscular VSD comes to the office with complaints from the mother of fatigue and poor feeding over the past month. You note the child has not gained weight since the previous visit 2 months ago. The child is apathetic, tachypneic, and has wheezes and crackles on lung auscultation.

D

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The Cardiovascular System

Answers

175. The answer is c. *(Hay et al, pp 339, 346-347, 349-352. Kleigman et al, pp 345-351. McMillan et al, pp 749, 751-754, 759-764. Rudolph et al, pp 360-361, 364-365, 367-368, 371.)* The clinical presentation is that of a tricyclic antidepressant ingestion. In smaller children the central nervous system (CNS) symptoms of drowsiness, lethargy, coma, and seizures are more commonly seen than the cardiac effects of tachycardia, initial hypertension followed by hypotension, and a variety of cardiac effects such as widening of the QRS complex and bundle branch blocks which are often seen in adolescents. Close monitoring in the ICU and tricyclic antidepressant Fab antibody fragments (if available) are administered.

Acetaminophen ingestion results initially in nausea, vomiting, and diaphoresis. These early symptoms often resolve in about 24 to 48 hours progressing to right upper quadrant abdominal pain and liver function enzyme elevation. By 2 to 3 days after the ingestion the peak liver function abnormalities are noted and by 4 days to about 2 weeks either recovery of liver function is noted or complete liver failure ensues. Therapy is with *N*-acetylcysteine. Narcotic ingestions result in respiratory depression as a major symptom during an acute ingestion as described; naloxone (repeated doses may be necessary) is the therapeutic choice. Ethanol is used to compete with methanol (not a pill); methanol ingestion causes mild inebriation, visual disturbances, nausea and vomiting, drowsiness, and profound acidosis. Iron overdose (treated with deferoxamine) leads to early nausea, vomiting, diarrhea, abdominal pain, bloody stools, hypotension, and ultimately gastric scarring.

176. The answer is b. *(Hay et al, pp 580-582. Kleigman et al, pp 1141-1145. McMillan et al, pp 1662-1669. Rudolph et al, pp 1901-1904.)* The diagnosis of rheumatic fever can be difficult because no single clinical manifestation or laboratory test is confirmatory. However, an accurate diagnosis must be made since treatment of the acute problems promptly and effectively is required and long-term antibiotic prophylaxis to prevent recurrences must be instituted.

To assist in diagnosis, the American Heart Association identified a set of major and minor standards relating to the manifestations of the disease, called the *Jones criteria* (modified), and recommends these criteria be applied in the diagnosis of every patient with possible rheumatic fever. The major criteria are carditis, arthritis, erythema marginatum, chorea, and subcutaneous nodules. The minor criteria are arthralgia (joint pain with no objective findings), fever or history of rheumatic fever, increased erythrocyte sedimentation rate (ESR), positive C-reactive protein, increased WBC and anemia, and prolonged PR and QT intervals on ECG. To make the diagnosis of rheumatic fever, the following criteria should be met: two major manifestations, one major and two minor manifestations plus strong evidence of a preceding group A β-hemolytic streptococcal infection (culture, rapid antigen-antibody rise, or elevation), or scarlet fever.

177. The answer is b. *(Hay et al, pp 570-572. Kleigman et al, pp 1906-1911. McMillan et al, pp 1547-1549. Rudolph et al, pp 1820-1823.)* This child has tetralogy of Fallot, which consists of right ventricular outflow obstruction (pulmonary stenosis), VSD, dextroposition of the aorta, and right ventricular hypertrophy. The radiograph shows the typical "boot-shaped" heart, while the ECG demonstrates the increased right ventricular forces. Children with tetralogy of Fallot may have cyanotic episodes ("tet spells") associated with acute reduction in pulmonary blood flow. Typically, these spells are self-limited, lasting no more than 30 minutes. Assuming the knee-chest position is thought to increase peripheral resistance, decreasing the amount of right-to-left shunting and thus increasing pulmonary blood flow. Alternative therapies include morphine sulfate and propranolol. Prolonged hypoxia can lead to acidosis; correction may require infusion of sodium bicarbonate.

Patent ductus arteriosus is likely to present in this aged child with a constant, machine-like murmur, and if the flow is big enough, with evidence of progressive congestive heart failure. An ASD will have a fixed split-second heart sound and a pulmonic stenosis murmur; it can also present with progressive evidence of heart failure if the flow across the ASD is large enough. Transposition of the great vessels with a patent foramen ovale and hypoplastic left heart syndrome are neonatal conditions; presentations are in the newborn period.

178. The answer is a. *(Hay et al, p 233. Kleigman et al, p 585. McMillan et al, p 2194. Rudolph et al, pp 696-698.)* Identification of those with a genetic predisposition to hypercholesterolemia and of the factors that increase the risk of the condition is recommended so that dietary and other

measures to reduce serum lipids can be introduced if indicated. Children with a first- or second-degree relative with early onset of coronary heart disease should be evaluated early in life but after 2 years of age. Other known risk factors include obesity, diabetes, hypertension, and smoking. No change in current infant dietary practice is recommended for children under 2 years of age. The high total fat content of the infant diet is considered to be biologically sound, given the need for lipid for a developing nervous system and the infant's limited capacity for an intake of high volume during this period of rapid growth. It is generally agreed that a dietary fat intake of 40% of calories is excessive over the age of 2 years. There is, however, concern about the potential loss of minerals such as iron, zinc, and calcium when dietary fat is reduced below 30% of calories in children 2 to 18 years of age. Principal sources of fat in the American diet are meat and milk, which are also sources of minerals such as iron and calcium. Simple modifications in the current US diet of children of these ages (trimming excess fat from meat and drinking 1% fat milk) would effect a reduction of fat intake by 5% of calories without the risk of lowering mineral intake. It would be helpful in this situation to determine whether the uncle had hypercholesterolemia. Yearly ECGs and lipid-lowering agents are not indicated in this situation.

179. The answer is d. *(Hay et al, p 931. Kleigman et al, pp 979-982. McMillan et al, pp 2413-2416. Rudolph et al, p 1196.)* The picture shows edema around the neck. This condition often worsens in adolescents. Although hereditary angioedema is relatively rare as a cause of edema, the recurrent episodes in late childhood, the normal laboratory results, and the family history make the other choices less likely. Hereditary angioedema, transmitted as an autosomal dominant trait, is a result of inadequate function (owing to either deficient quantity or quality) of an inhibitor of the first step in the complement cascade, which results in the excessive production of a vasoactive kinin. In addition to otherwise asymptomatic subcutaneous edema, edema can occur in the gastrointestinal tract and produce the symptoms mentioned in the question. Laryngeal edema with airway obstruction can also occur.

180. The answer is c. *(Hay et al, pp 569, 1032. Kleigman et al, pp 2386-2389. McMillan et al, p 2635-2336. Rudolph et al, pp 2087-2089.)* Short stature, neck webbing, sexual infantilism, and a shieldlike chest with widely spaced nipples are signs of Turner syndrome, which is usually associated with an XO karyotype. Aortic coarctation occurs in about 15% to 20% of those with this

disorder. Down syndrome is most commonly associated with endocardial cushion defects or VSD. Marfan syndrome (mutation 15q21.1) is associated with dilatation of the aorta and mitral and aortic regurgitation. Ellis-van Creveld syndrome (defect 4p16) is associated with ASD.

181. The answer is b. *(Kleigman et al, pp 1878-1881. McMillan et al, pp 335-338. Rudolph et al, p 1781.)* The incidence of congenital heart disease in the population is about 1%. The risk of congenital heart disease in a family with one child born with heart disease varies depending on the type of lesion the first child had but, overall, the rate averages from about 2% to 6% if the heart disease is not associated with a diagnosable syndrome. The risk of congenital heart disease for an infant is increased if there is a history of congenital heart disease in the mother. If two children with congenital heart disease are diagnosed, the risk escalates to 20% to 30%.

182. The answer is a. *(Kleigman et al, pp 1883-1885. McMillan et al, pp 287, 1329-1332, 1346-1350, 1354-1357, 1378-1380. Rudolph et al, pp 1788-1794, 1797-1798, 1820-1823.)* Most children with an ASD are asymptomatic; the lesion is found during a routine examination. In older children, exercise intolerance can be noted if the lesion is of significant size. On examination, the pulses are normal, a right ventricular systolic lift at the left sternal border is palpable, and a fixed splitting of the second heart sound is audible. For lesser degrees of ASD, surgical treatment is more controversial.

VSD commonly present as a harsh or blowing holosystolic murmur best heard along the left lower sternum, often with radiation throughout the precordium. Tricuspid regurgitation is a middiastolic rumble at the lower left sternal border; a history of birth asphyxia or findings of other cardiac lesions often is present. Tetralogy of Fallot is a common form of congenital heart disease. The four abnormalities include right ventricular outflow obstruction, VSD, dextroposition of the aorta, and right ventricular hypertrophy; the cyanosis presents in infants and in young children. Mitral valve prolapse occurs with the billowing into the atria of one or both mitral valve leaflets at the end of systole. It is a congenital abnormality that frequently manifests only during adolescence or later. It is more common in girls than in boys and seems to be inherited in an autosomal dominant fashion. On clinical examination, an apical murmur is noted late in systole, which can be preceded by a midsystolic click. The diagnosis is confirmed with an echocardiogram that shows prolapse of the mitral leaflets during mid to late systole.

183. The answer is c. *(Hay et al, pp 582-583. Kleigman et al, pp 1036-1042. McMillan et al, pp 1015-1023. Rudolph et al, pp 844-845.)* Many conditions can be associated with prolonged fever, a limp caused by arthralgia, exanthem, adenopathy, and pharyngitis. Conjunctivitis, however, is suggestive of Kawasaki disease. The fissured lips, although common in Kawasaki disease, could occur after a long period of fever from any cause if the child became dehydrated. The predominance of neutrophils and high ESR are common to all. An increase in platelets within this constellation of symptoms, however, is found typically in Kawasaki disease. Kawasaki disease presents as a picture of prolonged fever, rash, epidermal peeling on the hands and feet (especially around the fingertips), conjunctivitis, lymphadenopathy, fissured lips, oropharyngeal mucosal erythema, and arthralgia or arthritis. The diagnosis is still possible in the absence of one or two of these physical findings. Coronary artery aneurysms can develop, as can aneurysms in other areas. Initial treatment is typically intravenous immunoglobulin (IVIG) and high-dose aspirin. The child will usually defervesce shortly after the infusion. Aspirin is typically kept at a higher dose until the platelet count begins to decrease, and then is continued at a lower dose for several weeks. While bacterial infection is in the differential diagnosis for this patient's presentation and blood cultures are usually part of the evaluation, IV vancomycin should be reserved for a culture-proven susceptible organism resistant to other antibiotics, or as empiric therapy in a critically ill patient.

184. The answer is a. *(Hay et al, pp 587-588. Kleigman et al, pp 1970-1971. McMillan et al, pp 1614-1624. Rudolph et al, pp 1860-1861.)* The findings of pallor, dyspnea, tachypnea, tachycardia, and cardiomegaly are common in congestive heart failure regardless of the cause. The most common causes of myocarditis include adenovirus and coxsackievirus B, although many other viruses can cause this condition. The constellation of findings in the question point to myocarditis as the etiology of this patient's condition.

The echocardiogram in the question shows ventricular and left atrial dilatation as well as poor ventricular function. With glycogen storage disease of the heart muscle thickening would be expected and with pericarditis a pericardial effusion would be seen. The echocardiogram findings in this case are not consistent with an aberrant origin of the left coronary artery, although the origin of the coronary arteries can be difficult to visualize. On ECG, the voltages of the ventricular complexes seen with aberrant origin of the left coronary artery are not diminished, and a pattern of myocardial infarction can be seen. Voltages from the left ventricle are usually high in

endocardial fibroelastosis, and both right and left ventricular forces are high in glycogen storage disease of the heart.

185. The answer is e. *(Hay et al, pp 597-600. Kleigman et al, pp 1945-1947. McMillan et al, pp 1679-1681. Rudolph et al, pp 1854-1856.)* Congestive heart failure from any cause can result in mild cyanosis, even in the absence of a right-to-left shunt, and in poor peripheral pulses when cardiac output is low. Congestive heart failure from many causes can be associated with a rapid pulse rate (up to 200 beats per minute). A pulse rate greater than 250 beats per minute, however, should suggest the presence of a tachyarrhythmia. Common causes for supraventricular tachycardia include Wolff-Parkinson-White (WPW) syndrome, congenital heart disease, and sympathomimetic drugs. In this patient, evaluation for WPW syndrome and cardiac abnormalities must be accomplished after the congestive heart failure from the increased heart rate is under control.

186. The answer is c. *(Hay et al, pp 597-600. Kleigman et al, pp 1945-1947. McMillan et al, pp 1679-1681. Rudolph et al, pp 1854-1856.)* Supraventricular tachycardia, as shown on the ECG, is characterized by rapid heart rate (in this case about 250 beats per minute), little rate variability, and a consistent P wave for each QRS complex. Prolonged SVT can lead to heart failure with hepatomegaly and respiratory compromise. Fetal SVT can lead to hydrops fetalis. The first-line treatment is to stimulate the vagus nerve using techniques such as carotid massage, immersion of the face in cold water, or voluntary straining. Rapid infusion of IV adenosine can affect resolution if the maneuvers are not successful. Verapamil is contraindicated in this age group, as it may cause acute hypotension and cardiac arrest. Synchronized DC cardioversion may be performed in patients in shock or with heart failure; it must, however, be synchronized to the QRS complex. Transthoracic pacing is typically useful in bradyarrhythmias. A precordial thump might help individuals with an acute arrest from ventricular fibrillation, but does not resolve SVT.

187. The answer is d. *(Hay et al, pp 563-564. Kleigman et al, pp 1915-1918. Rudolph et al, pp 1817-1818.)* A quadruple rhythm associated with the murmur of tricuspid regurgitation and a middiastolic murmur at the lower left sternum suggests the diagnosis of Ebstein anomaly (downward displacement of the tricuspid valve). The presence of right atrial hypertrophy and right ventricular conduction defects confirms the diagnosis. Both tricuspid regurgitation with pulmonic stenosis and tetralogy of Fallot give ECG evidence of

right ventricular enlargement. The WPW syndrome, which frequently accompanies Ebstein malformation, is not associated with murmurs or cyanosis as an isolated entity, but also includes supraventricular tachycardia.

188. The answer is c. *(Hay et al, p 598. Kleigman et al, p 1945-1947. McMillan et al, p 1677. Rudolph et al, pp 1452-1453.)* The child described in the question has no cyanosis or murmur, no cardiac or pulmonary vascular abnormalities by chest x-ray, and no evidence of structural anomalies by echocardiogram, and therefore is unlikely to have an underlying gross anatomic defect. The ECG pattern in the figure shows the configuration of preexcitation, the pattern seen in the WPW syndrome. These patients have an aberrant atrioventricular conduction pathway, which causes the early ventricular depolarization appearing on the ECG as a shortened PR interval. The initial slow ventricular depolarization wave is referred to as the *delta wave*. Seventy percent of patients with WPW syndrome have single or repeated episodes of paroxysmal supraventricular tachycardia, which can cause the symptoms described in the question. The preexcitation ECG pattern and WPW syndrome can occur in Ebstein malformation, but this is unlikely in the absence of cyanosis and with a normal echocardiogram. If ventricular tachycardia were present, the symptoms would likely be more profound. Active play and exposure to over-the-counter medications containing sympathomimetics in a healthy 4-year-old child can cause symptoms such as those described in the question in children with WPW syndrome by precipitating paroxysmal supraventricular tachycardia.

189. The answer is b. *(Hay et al, pp 822-824. Kleigman et al, pp 1001-1010. McMillan et al, pp 2538-2543. Rudolph et al, pp 836-839.)* Juvenile rheumatoid arthritis (JRA, or Still disease) frequently causes spindle-shaped swelling of finger joints and can involve unusual joints such as the sternoclavicular joint. Presentation of JRA occurs as either polyarthritis (five or more joints, systemic symptoms not so severe or persistent), pauciarticular (four or fewer joints, lower-extremity joints, extra-articular disease unusual), or systemic disease (severe constitutional disease, systemic symptoms prior to arthritis, rheumatoid rash, high spiking fevers, variable joint involvement). This disorder can be associated with spiking high fevers and diffuse rash, which are not a feature of rheumatic fever, toxic synovitis, or osteoarthritis. Although septic arthritis can affect any joint, it would not be likely to affect finger joints by causing spindle-shaped swellings. Toxic synovitis usually involves larger joints, such as the hip, and osteoarthritis is not a disease of childhood.

190. The answer is c. *(Hay et al, pp 573-574. Kleigman et al, pp 1913-1915. McMillan et al, pp 1543-1546. Rudolph et al, pp 1816-1817.)* Patients with tricuspid atresia typically have a hypoplastic right ventricle, and therefore the ECG shows left-axis deviation and left ventricular hypertrophy; this translates to a left ventricular impulse on physical examination. Almost all other forms of cyanotic congenital heart disease are associated with elevated pressures in the right ventricle and increased right ventricular impulse. In those conditions, therefore, the ECG will show right-axis deviation and right ventricular hypertrophy.

191. The answer is b. *(Hay et al, pp 575-577. Kleigman et al, pp 1918-1922. McMillan et al, pp 1537-1539. Rudolph et al, pp 1823-1826.)* Transposition of the great vessels with an intact ventricular septum presents with early cyanosis, a normal-sized heart (classic "egg on a string" radiographic pattern in one-third of cases), normal or slightly increased pulmonary vascular markings, and an ECG showing right-axis deviation and right ventricular hypertrophy. In tetralogy of Fallot, cyanosis is often not seen in the first few days of life. Tricuspid atresia, a cause of early cyanosis, causes diminished pulmonary arterial blood flow; the pulmonary fields on x-ray demonstrate a diminution of pulmonary vascularity, and left-axis and left ventricular hypertrophy are shown by ECG. Total anomalous pulmonary venous return below the diaphragm is associated with obstruction to pulmonary venous return and a classic radiographic finding of marked, fluffy-appearing venous congestion ("snowman"). In pulmonic atresia with an intact ventricular septum, cyanosis appears early, the lung markings are normal to diminished, and the heart is large.

192. The answer is d. *(Hay et al, pp 567-568. Kleigman et al, p 1905. McMillan et al, pp 1603-1605. Rudolph et al, pp 1797-1798.)* Mitral valve prolapse occurs with the billowing into the atria of one or both mitral valve leaflets at the end of systole. It is a congenital abnormality that frequently manifests only during adolescence or later. It is more common in girls than in boys and seems to be inherited in an autosomal dominant fashion. On clinical examination, an apical murmur is noted late in systole, which can be preceded by a click. The diagnosis is confirmed with an echocardiogram that shows prolapse of the mitral leaflets during mid to late systole. The ECG and chest x-ray are usually normal. β-Blockers and digitalis are unlikely to be required, and penicillin prophylaxis for dental procedures for patients with mitral valve prolapse is no longer recommended by the American Heart Association.

193. The answer is d. *(Hay et al, pp 572-573, 575-577. Kleigman et al, pp 1912-1913, 1915. McMillan et al, p 355. Rudolph et al, pp 1818-1820.)* The vignette describes an infant with a ductal-dependant cyanotic congenital heart lesion. In this example, the child had pulmonary atresia without a corresponding VSD; another example would have been transposition of the great vessels without a septal defect to allow mixing of oxygenated and nonoxygenated blood. Cyanotic infants who do not improve their saturations with supplemental oxygen should be evaluated carefully for structural heart disease. The ductus arteriosis typically closes in the first few hours of life; thus, these children will develop their cyanosis in the same time frame. Prostaglandin E_1 will help keep the ductus patent until a surgical procedure can be performed. PGE_1 does have the tendency to cause hypoventilation, so arrangements must be made for a potential artificial airway if necessary. Indomethacin closes a patent ductus arteriosis. Adenosine is used for supraventricular tachycardia.

194 to 196. The answers are 194-b, 195-d, 196-c. *(Hay et al, pp 567-568, 861, 871-872. Kleigman et al, pp 514, 2085, 2717-1718. McMillan et al, pp 2645, 2650, 2656. Rudolph et al, pp 738, 764-768, 2089.)* Thrombocytopenia-absent radius syndrome (TAR) is diagnosed in the newborn who demonstrates profound thrombocytopenia, bilateral absence of radius, and abnormally shaped thumbs. Cardiac lesions include TOF and ASD. About 40% of patients die in the newborn period as a result of low-platelet-induced bleeding.

Noonan syndrome, the "male Turner syndrome," occurs in both sexes. The most common features include short stature, downslanting palpebral fissures, ptosis, low set and malformed ears, webbed neck, shieldlike chest, pulmonic stenosis, and cryptorchidism. Mental retardation is seen in one-forth of affected individuals. It is associated with advanced paternal age.

Mitral valve prolapse is seen in patients with Ehlers-Danlos syndrome. Patients with Ehlers-Danlos syndrome have hyperextensibility and easy bruising, joint hypermobility (leading to joint dislocations and sprains), skin that is velvety to touch, and tissue fragility. Six different kinds are described with "classic" and "hypermobility" forms predominating.

197 to 200. The answers are 197-b, 198-c, 199-a, 200-d. *(Hay et al, pp 556-558, 585-586. Kleigman et al, pp 1019, 1888-1891, 1967-1969, 2477. McMillan et al, pp 466-467, 1575-1578, 1638-1641, 1681-1682. Rudolph et al, pp 847, 1858, 1860-1862, 1869-1877.)* Hypertrophic cardiomyopathy (HCM),

also called idiopathic hypertrophic subaortic stenosis, is often inherited in autosomal dominant fashion with variable penetrance. Many children are completely asymptomatic, while others may have complaints of weakness, fatigue, chest pain, and syncope. A midsystolic ejection murmur and a left ventricular lift sometimes can be appreciated on physical examination. The diagnosis is confirmed by echocardiography. HCM can be a cause of arrhythmia and death in young athletes.

Prolonged QT syndrome occurs in 1:10,000 to 1:15,000 children, usually first causing syncopal episodes in late childhood or adolescence. During the syncope episode arrhythmias may be noted, including ventricular fibrillation. These episodes may result in death. QT intervals are elongated on ECG. An autosomal recessive form associated with deafness (Jervell-Lange-Nielsen syndrome) and an autosomal dominant form (Romano-Ward syndrome) has been described.

Neonatal lupus is a rare manifestation of transferred maternal IgG autoantibodies. Infants can have thrombocytopenia, neutropenia, rash, liver dysfunction, and a congenital heart block. Most manifestations are self-resolved; however, the congenital heart block is permanent and frequently requires pacing.

Congestive heart failure can result from a number of causes, including congenital heart disease, Kawasaki disease, metabolic cardiomyopathies, arrhythmias, and viral myocarditis. The case presented is typical of an infant with heart failure. Such infants are weak, diaphoretic, have poor weight gain, and may be tachypneic with retractions. Lung findings can include crackles, wheezes, or both. Prompt evaluation by a pediatric cardiologist is imperative.

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